Fixed subaortic stenosis after repair of ostium primum defects

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SUMMARY   Three patients who had closure of an ostium primum defect developed severe fixed subaortic stenosis requiring surgical removal three to 11 years later.

In two, the basic anatomy of the attachment of the superior portion of the anterior mitral cusp and submitral apparatus predisposed to the formation or deterioration of this complication. Both had sub-valvar gradients and angiographic narrowing of the outflow tract in systole and diastole. In the third patient the strut of the Hancock prosthesis used to replace a double orifice mitral valve impinged on the outflow tract to stimulate the formation of a serious fixed obstruction.

Subaortic gradients before operation must be specifically looked for, particularly in those with the characteristic radiological deformity of the submitral apparatus, and at operation the subaortic region must be inspected.

Postoperatively the search for developing subaortic stenosis must include regular M-mode and 2-dimensional echocardiography, and cardiac catheterisation may be required.

Although a rare complication, subaortic stenosis is a progressive and damaging lesion which requires early recognition and treatment despite the apparent well-being of the patient.

Since 1954 when the first successful repair of an ostium primum defect was reported,[1] increased experience and improved techniques have lowered the mortality and morbidity of the surgical treatment of this lesion,[2-5] but residual mitral regurgitation is common and may be severe enough to require reoperation[5-9] and atrial rhythm disorders or varying degrees of heart block may be precipitated prematurely or deteriorate.[2-5] The late development of fixed subaortic stenosis after repair of ostium primum defects, however, is less well recognised and the purpose of this paper is to describe three patients who developed this. The pathogenesis and the need for early recognition and treatment of this condition is discussed.

Patients

CASE 1

Investigation of this boy at the age of 4 years confirmed the clinical diagnosis of an ostium primum defect. A left-to-right shunt of 2:1 was found, with a normal pulmonary artery pressure and

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moderate mitral regurgitation. A subvalvar peak systolic gradient of 15 mmHg was found between the left ventricle and aorta, and was ignored. Left ventricular angiography in the anteroposterior view showed the classical "goose neck" appearance of the left ventricular outflow tract in diastole, but with narrowing also in systole (Fig. 1A and B). Review of these old films showed that slight narrowing of the outflow tract could be seen on the lateral views in systole and diastole (Fig. 1C).

Operation through the right atrium by Mr Keith Ross confirmed the standard sized ostium primum defect, with a complete cleft in the anterior leaflet of the mitral valve. At the apex of the cleft the cusp tissue was deficient and was tethered by short chordae inserting into the ventricular septum. The cleft was repaired by three interrupted sutures and the atrial septal defect closed with a pericardial patch.

The patient escaped routine follow-up at the National Heart Hospital as he lived in Scotland, but four years later he became breathless and presented with dyspnoea, electrocardiographic left ventricular hypertrophy, and cardiomegaly on the chest radiograph. At this time there was a harsh ejection systolic murmur with a carotid thrill, a jerky pulse, and a soft immediate aortic diastolic murmur, suggesting
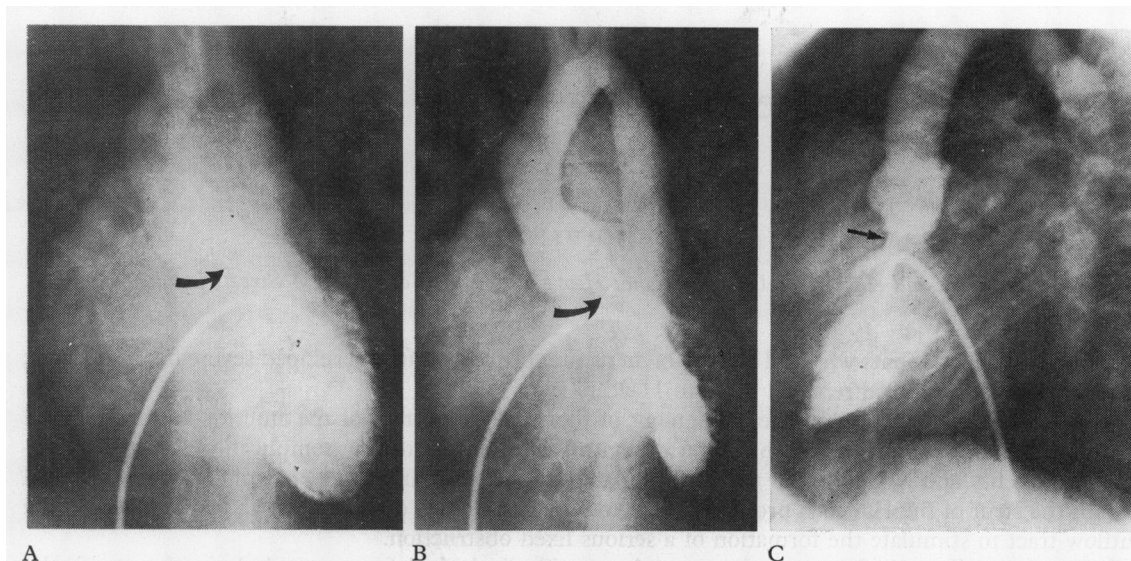
Fig. 1 *Preoperative left ventricular angiograms in case 1 showing narrowing of the left ventricular outflow tract both in systole and diastole (arrowed). (A) Diastolic frame, anteroposterior (AP) view; (B) systolic frame, AP view; (C) systolic frame, lateral view.*

the diagnosis of fixed subaortic obstruction. This was confirmed by 2-dimensional echocardiography (Fig. 2A). M-mode echocardiography showed early systolic closure of the aortic valve, and a scan from the aorta to the left ventricle suggested a long fixed narrowing of the outflow tract. The resting pulmonary artery pressure was normal and there was no residual left-to-right shunt but there was a resting peak systolic gradient of 60 mmHg beneath the aortic valve with angiographic confirmation of a fixed subaortic obstruction (Fig. 2B).

At reoperation a complete fibrous ring was found 2 mm beneath the aortic valve, attached to the abnormally positioned superior portion of the anterior cusp of the mitral valve and to the under-surface of the right coronary cusp of the aortic valve. In addition, the short abnormal chordae and aberrant papillary muscles further contributed to the narrowing. The fibrous tissue was excised, almost completely relieving the obstruction, but the subaortic region still appeared to be narrow because of the abnormal attachments of the mitral valve and this could not be relieved without disturbing mitral function.

Reinvestigation one month after operation showed a resting peak systolic gradient of only 12 mmHg across the left ventricular outflow tract. Twenty-three months later, though the patient remained well, he had an M-mode echocardiogram performed during routine follow-up in Scotland and this showed early systolic closure of the aortic valve

together with a narrowed left ventricular outflow tract. We were concerned that important subaortic stenosis had recurred. When seen at the National Heart Hospital he had once more a harsh ejection systolic murmur at the base and an immediate diastolic murmur together with jerky carotid pulses and a carotid thrill suggesting renewed subaortic stenosis. Two-dimensional echocardiography confirmed the presence of fixed subaortic stenosis, and a resting gradient of 60 mmHg rising to 80 mmHg with isoprenaline was found at catheterisation. Left ventricular angiography confirmed fixed subaortic obstruction with the same appearances as before the second operation. As the electrocardiogram had shown improvement in comparison with preoperative records and the child was asymptomatic, it was decided to observe him until some evidence of deterioration occurred. This unusual policy was recommended because we considered that the mitral valve and papillary muscles would probably need to be removed in order to relieve the obstruction and we hope that by the time this becomes necessary improved mitral valve protheses will be available.

CASE 2
This boy had closure of an ostium primum defect performed at the age of 10 years in another centre abroad; he also had a double orifice mitral valve replaced by a 2·5 cm Hancock prosthesis. Before operation he had had a 3:1 left-to-right shunt with a normal pulmonary artery pressure, and left ven-
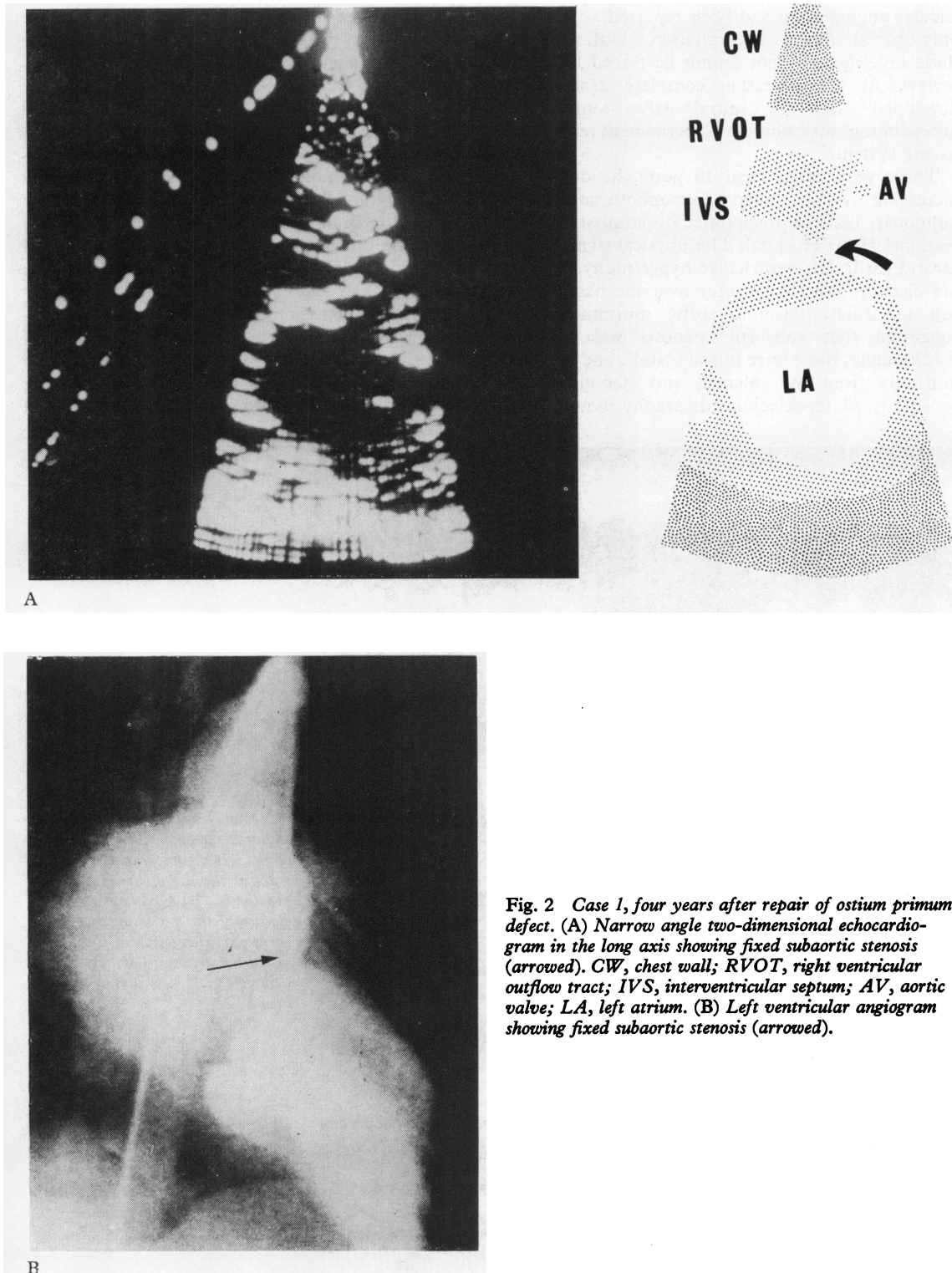
Fig. 2 *Case 1, four years after repair of ostium primum defect. (A) Narrow angle two-dimensional echocardiogram in the long axis showing fixed subaortic stenosis (arrowed). CW, chest wall; RVOT, right ventricular outflow tract; IVS, interventricular septum; AV, aortic valve; LA, left atrium. (B) Left ventricular angiogram showing fixed subaortic stenosis (arrowed).*

tricular angiography had been reported as showing only trivial mitral regurgitation. Unfortunately these investigative data cannot be traced for us to review. At the operation complete heart block developed after the mitral valve replacement necessitating insertion of a permanent epicardial pacing system.

Three years later, aged 13 years, he developed increasing dyspnoea over a four-month period and pulmonary oedema precipitated his admission to the National Heart Hospital. The physical signs were a carotid thrill, left ventricular hypertrophy, a grade 3/6 ejection systolic murmur over the base, and a soft immediate diastolic aortic murmur. These suggested that subaortic stenosis was present. Additionally, there were mitral systolic and diastolic murmurs from the calcified and stenotic mitral prosthesis. M-mode echocardiography showed early

systolic closure of the aortic valve and severe narrowing of the left ventricular outflow tract suggestive of significant subaortic stenosis as well as severe mitral stenosis (Fig. 3). Emergency cardiac catheterisation showed a peak systolic gradient of 70 mmHg across the left ventricular outflow tract and a 25 mmHg gradient across the stenotic, calcified prosthesis. The pulmonary artery pressure was 100/60 mmHg, with no left-to-right shunt. Left ventricular angiography showed a thickened left ventricle; the ring of the prosthesis appeared to impinge on, and to be causing narrowing of the left ventricular outflow tract in systole and diastole (Fig. 4).

Soon after admission the boy developed ventricular fibrillation in the ward during an attack of pulmonary oedema. At emergency operation by Mr Magdi Yacoub, an irregular fibrous subaortic ring was found to have formed over the suture material
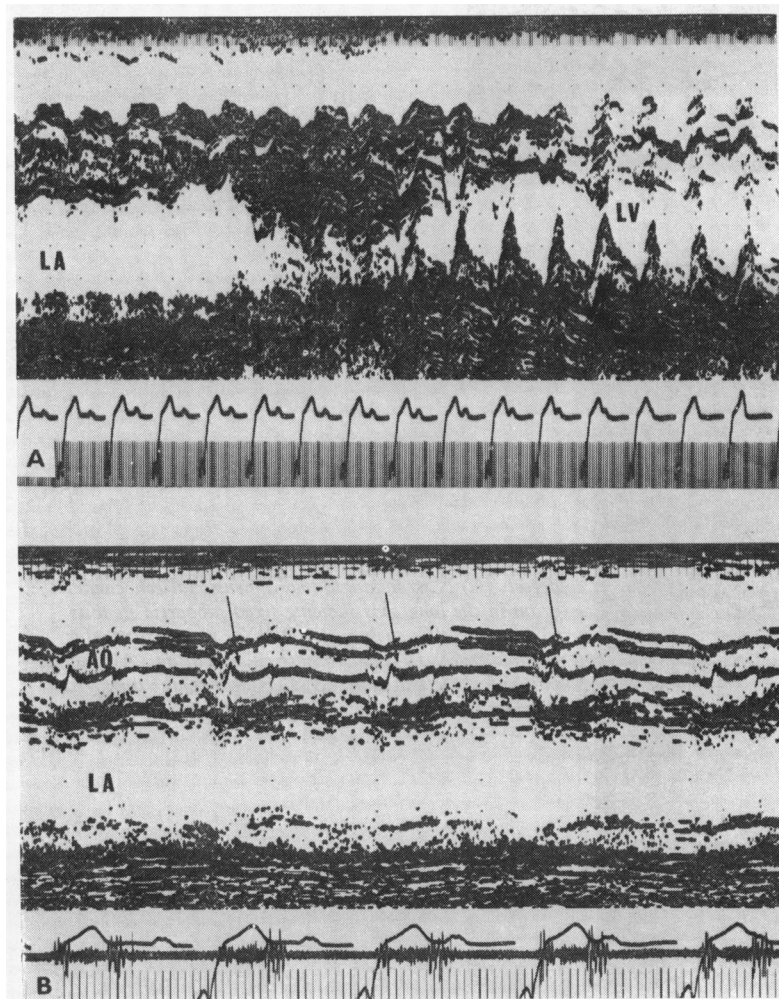

Fig. 3 *M-mode echocardiogram in case 2, three years after closure of an ostium primum defect and mitral valve replacement with a Hancock prothesis. (A) Scan from the left ventricle to aorta showing severe narrowing of the left ventricular outflow tract at the site of the Hancock prosthesis. (B) Early systolic closure of the aortic valve (arrowed) suggestive of fixed subaortic stenosis. LV, left ventricle; LA, left atrium; AO, aorta.*
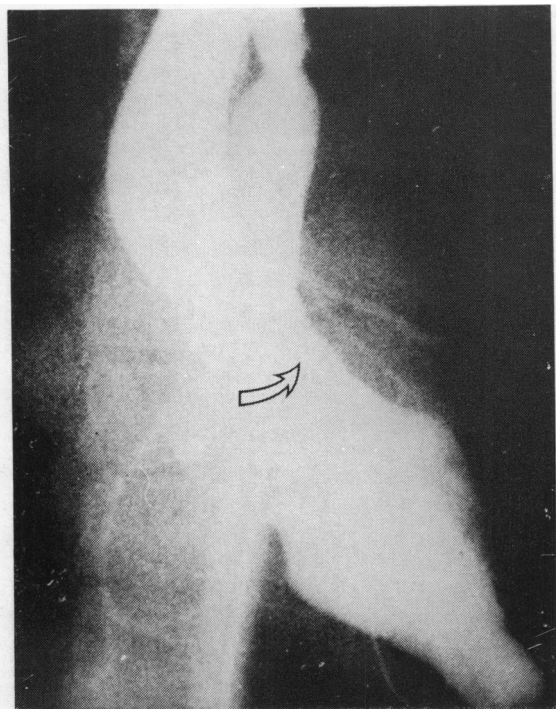
Fig. 4 *Systolic frame of left ventricular angiogram in case 2, three years after closure of ostium primum defect and mitral valve replacement with Hancock prosthesis showing fixed subaortic stenosis caused by impingement of the strut and fibrous tissue deposition into the left ventricular outflow tract (arrowed).*

left ventricle. Left ventricular angiography in the anteroposterior projection showed gross mitral regurgitation with abnormal attachment of the anterior mitral cusp, suggesting an ostium primum defect. Retrospectively the left ventricular outflow tract appeared narrowed both in systole and diastole but the area was obscured because in the anteroposterior projection it was overlapped by dense left atrial opacification (Fig. 5).
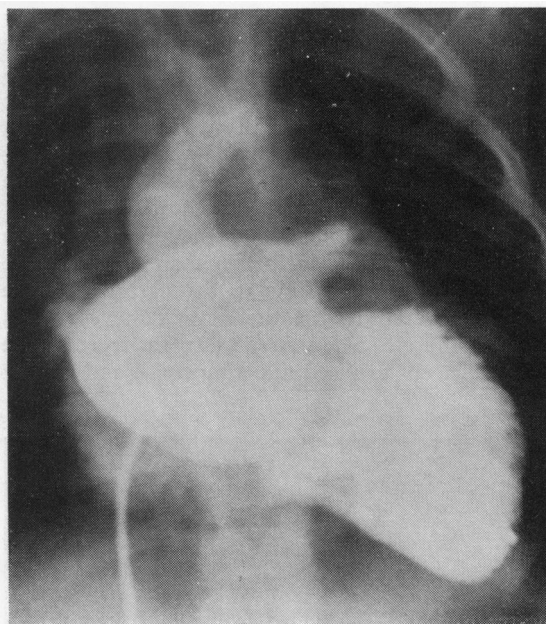

Fig. 5 *Preoperative left ventricular angiogram in case 3, showing gross mitral regurgitation and narrowing of the left ventricular outflow tract.*

incorporated in the ventricular septum and was attached also to the rim of the Hancock prosthesis which protruded into the left ventricular outflow tract beneath the aortic valve. This was dissected out and in so doing a ventricular septal defect was produced, and patched. A fresh inverted 2·6 cm aortic homograft valve mounted on a Dacron cylinder was used to replace the mitral prosthesis. The boy died despite prolonged assisted bypass and massive catecholamine support.

At necropsy by Dr E Olsen there appeared to be residual fibrous tissue causing left ventricular outflow tract obstruction which was contributed to by the Dacron stent supporting the mitral homograft. The heart is unfortunately no longer available for more detailed study.

CASE 3

At the age of 3 years this girl presented with severe mitral regurgitation and a small left-to-right shunt of 1·5:1 at atrial level. The pulmonary artery pressure was 27/10 mmHg and there was a peak systolic gradient of 21 mmHg between the aorta and

At operation by Mr Donald Ross, an unusually small ostium primum defect was found through which a complete cleft in the anterior leaflet of the mitral valve was sutured. After pericardial patch closure of the small primum defect the subaortic region was inspected through an aortotomy because of the preoperative finding of a subaortic gradient. Beneath the normal aortic valve aberrant papillomatous mitral cusp tissue attached directly to the ventricular septum in the region of the membranous septum was found and excised and the area was oversewn.

After the operation the patient attended another clinic and remained asymptomatic. Eleven years later, however, now aged 14, she developed severe left ventricular hypertrophy together with jerky carotid pulses, a carotid thrill, a loud praecordial ejection systolic murmur and an immediate aortic

diastolic murmur. In retrospect, a long ejection systolic murmur had been present at the base and apex since operation and had been attributed to mitral regurgitation. M-mode echocardiography showed early systolic closure of the aortic valve and a narrowed left ventricular outflow tract. Two-dimensional echocardiography showed classic fixed subaortic stenosis (Fig. 6A). Severe subaortic stenosis was confirmed at catheterisation by a resting subaortic peak systolic gradient of 120 mmHg. There was no left-to-right shunt and the pulmonary artery pressure was normal. Angiography confirmed severe subaortic obstruction and moderate aortic regurgitation (Fig. 6B).

At reoperation 11 years after the first procedure, a mass of subaortic fibrous tissue was found forming an obstruction incorporating previous sutures in the ventricular septum and opposite in the sutured cleft of the anterior mitral cusp, the apex of which appeared to be attached further than usual into the outflow tract contributing to the narrowing. The densely adherent fibrous tissue was removed with a layer of muscle. The mitral valve was freed at the angle adjacent to the conducting tissue and freeing
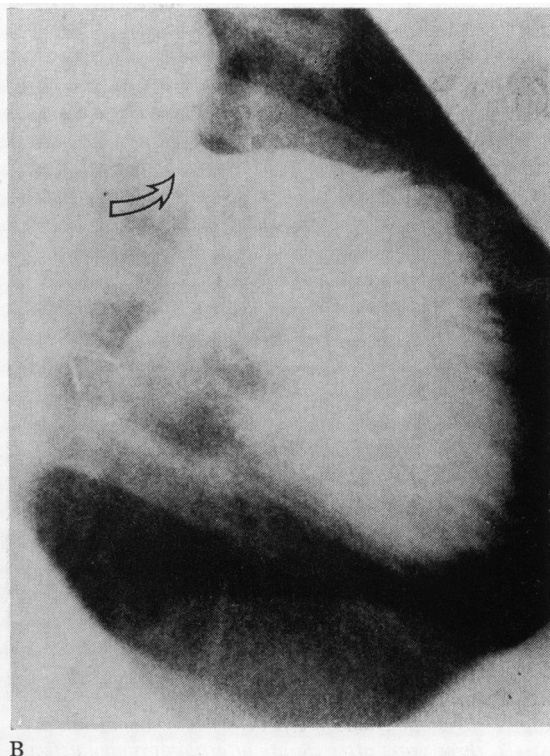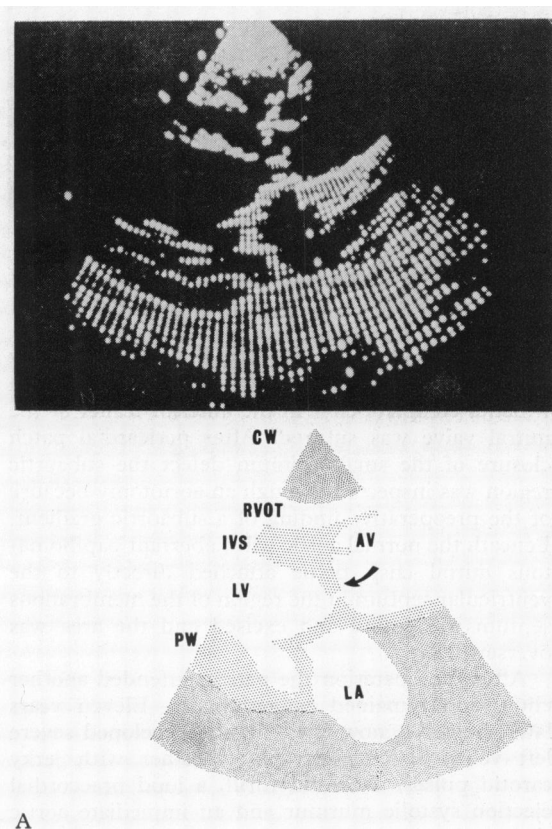

Fig. 6 *Investigations in case 3, 11 years after repair of ostium primum defect. (A) Wide angle two-dimensional echocardiogram in the long axis view showing fixed subaortic stenosis (arrowed). CW, chest wall; RVOT, right ventricular outflow tract; IVS, interventricular septum; AV, aortic valve; LV, left ventricle; PW, posterior left ventricular wall; LA; left atrium. (B) Left ventricular angiogram showing severe fixed subaortic stenosis (arrowed).*

the sutures produced a small iatrogenic ventricular septal defect which was closed. The ventricular septum was thick and a myomectomy was done. After removal of all this tissue the aortic valve became severely regurgitant from loss of support to the cusps and so it was excised and replaced with a 23 mm Carpentier Edwards xenograft.

After operation left ventricular failure recurred. Two further operations for persistence of the ventricular septal defect, and paraprosthetic aortic regurgitation, secondary to infection by a resistant *Staphylococcus epidermis*, were performed. At the last operation the xenograft was excised and a formal aortic root and valve replacement together with reimplantation of the coronary arteries was undertaken.[10]

Six months later she was well, free of cardiac

failure, with a competent aortic homograft and no clinical signs of subaortic obstruction. The heart size was considerably reduced on the chest radiograph and both M-mode and two-dimensional echocardiography showed no evidence of subaortic stenosis (Fig. 7).

Discussion

In view of the abnormal attachment of the anterior cusp of the mitral valve and the increased length of the left ventricular outflow tract, which are characteristics of atrioventricular defects,[11][12] it is perhaps surprising that organic subaortic stenosis is not found more frequently both before and after operations for both ostium primum defects and common atrioventricular canal. In these three patients the fixed subaortic stenosis was composed of the same tissue found in lone subaortic stenosis[13] and appeared to have formed in two because the attachments of the superior portion of the anterior mitral cusp appeared to narrow the outflow when the cleft was closed, and in the third because of impingement of the semi-rigid strut of the mitral prosthesis into the outflow tract. Unfortunately we do not know the exact anatomical angiographic details in the last

child whose double orifice mitral valve was unfortunately replaced, but in the first two it was possible to recognise the "potential" for the development of fixed subaortic obstruction. The striking feature before the first uncomplicated closure of the defects in cases 1 and 3 was the appearance of narrowing of the left ventricular outflow tract in systole as well as diastole. This appeared to be related to two factors, namely, unusual encroachment of the superior portion of the anterior mitral cusp into the outflow tract and also a short abnormal attachment of an aberrant papillary muscle appearing as a small cephalad filling defect in the outflow tract during systolic contraction. In both patients there was a small outflow tract gradient which was ignored in case 1.

Little attention is paid to subaortic stenosis in the preoperative investigation of ostium primum defects. Any gradient which might be present is rarely measured as the left ventricular angiogram is usually performed through a catheter passed via the atrial septal defect. We now recommend that left ventricular and systemic arterial pressures are measured simultaneously and routinely, especially where there is the particular appearance of left ventricular outflow tract narrowing in the systolic
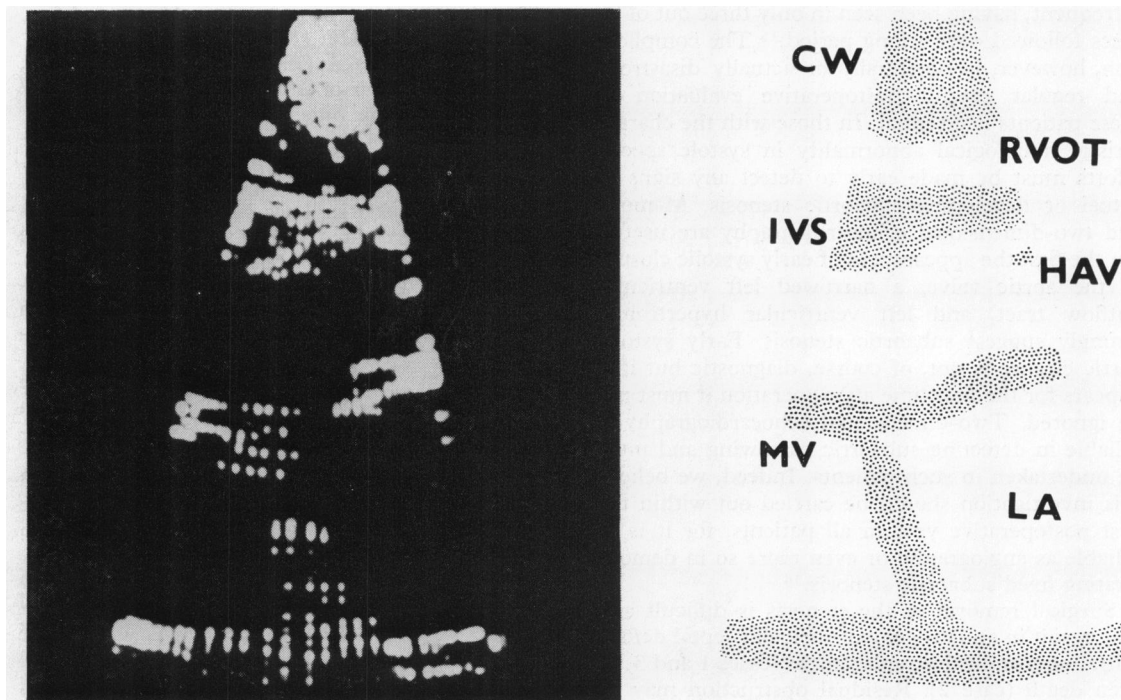

Fig. 7 *Narrow angle, long axial two-dimensional echocardiogram in case 3 after total aortic root and valve replacement showing the subaortic stenosis to have been completely relieved. CW, chest wall; RVOT, right ventricular outflow tract; IVS, interventricular septum; HAV, homograft aortic valve; MV, mitral valve; LA, left atrium.*

film of the left ventricular angiogram. Whether there is more muscle causing partial discontinuity between aortic valve and mitral valve in some of these patients, which might explain the subaortic stenosis, remains to be seen. The left ventricular outflow tract in these patients seems even longer than usual for an atrioventricular defect.

As a result of our experience we consider it mandatory to direct attention to the subvalvar region after closure of the primum defect and suturing of the cleft in patients with a small preoperative gradient and systolic narrowing on angiography. After surgical repair the left ventricular outflow tract is known to remain narrow in a number of cases,[9] [14] and we believe that once mitral regurgitation is improved and the atrial septal defect closed, forward flow is increased, resulting in increased turbulence in the already narrowed subvalvar area. This will stimulate the deposition of fibrous tissue and the narrowing will progress if left behind. We suggest that whenever the mitral valve is repaired or replaced in ostium primum defects, the surgeon specifically looks for a left outflow gradient and inspects the subaortic area through an aortotomy if one is found.

The development of fixed subaortic stenosis after repair of ostium primum defects appears to be infrequent, having been seen in only three out of 99 cases followed over a long period.[15] The complication, however, is potentially or actually disastrous and regular careful postoperative evaluation in these patients is essential. In those with the characteristic radiological abnormality in systole special efforts must be made early to detect any signs of actual or developing subaortic stenosis. M-mode and two-dimensional echocardiography are useful. On the first the appearances of early systolic closure of the aortic valve, a narrowed left ventricular outflow tract, and left ventricular hypertrophy strongly suggest subaortic stenosis. Early systolic aortic closure is not, of course, diagnostic but if it appears for the first time after operation it must not be ignored. Two-dimensional echocardiography is reliable in detecting subaortic narrowing and must be undertaken in such patients. Indeed, we believe this investigation should be carried out within the first postoperative year in all patients, for it is as reliable as angiography or even more so in demonstrating fixed subaortic stenosis.[16]

Surgical removal of the stenosis is difficult and may result in an iatrogenic ventricular septal defect, mitral or aortic valve replacement (cases 1 and 3) or even death (case 2). Residual obstruction may be left or may recur, as it did in case 1.

Thus, fixed subaortic stenosis appears to be a rare complication after repair of ostium primum

defects, but may be underdiagnosed. It must be thought of, investigated, and treated early despite the apparent well-being of the patient if the function of the left ventricle is to be preserved.

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